Benign Small Bowel Tumor

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The clinical record and histologic sections of 84 cases of benign small bowel tumor are reviewed. Manifestations of systemic diseases, congenital anomalies, and lesions of either the ileocecal valve or periampullary region were excluded. In the same time span there were 96 small bowel malignancies. Clinical presentation, pathologic findings, management and result are compared to the collected published experience of about 2000 cases. There were 36 leiomyomas, 22 lipomas, 9 angiomas, 6 neurofibromas and 4 fibromas. Thirty-six men and 48 women were affected; the majority in their fifth and sixth decade. Seventy-eight were operative and 6 autopsy diagnoses. The most common symptom was obstruction (42%) followed by hemorrhage (34%) and pain (22%), relative frequency differing for the various specific tumors. There were rarely significant physical findings. A diagnosis of small bowel tumor was made radiologically in 30 patients. Because of the nonspecificity of other signs and symptoms, an acute awareness of the possibility of small bowel tumor is mandatory for preoperative anticipation of the diagnosis. Local resection was performed in all with no deaths or significant postoperative complications.

BENIGN TUMORS are quite rare in the small bowel[15] and these lesions usually become clinically apparent only when they produce complications such as hemorrhage or obstruction.

Since the first collective review of benign small bowel tumors by Hearteaux[11] in 1899, information about the incidence and behaviour of these tumors has accrued slowly. The most recent extensive study of the literature was of 1340 cases by River *et al.*[16] in 1955.

The records of The New York Hospital-Cornell Medical Center were reviewed for the period between 1932 and 1972. There were 180 cases of primary tumors of the small bowel. Eighty-four of these were benign, including seven previously reported by Eckel[8] in 1948. Manifestations of systemic diseases, such as Peutz-Jegher's syndrome and Von Recklinhausen's disease were excluded as were ectopic pancreas, periampullary tumors, and ileococecal valve tumors. All carcinoid tumors were considered to be malignant. Six types of benign tumors were found; leiomyomas, lipomas, angiomas, adenomas, neurofibromas, and fibromas. Their anatomic distribution is given in Table 1.

This communication reports and analyses these 84 cases. Cumulative data from the collective review of River *et al.* and 657 cases[1,2,4-7,9,12-14,17,19,21,24] from major reviews since 1955 are tabulated, presented, and compared with our experience.

Clinical Presentation

Thirty-six of the tumors appeared in male patients and 48 in females, similar to a near equal sex distribution in reported series.[1,2,5,9,12,16] Thirty-one per cent were in their sixth and 28% in their fifth decade, not notably different from the age range of other series.[1,2,7,9,12,14,16] Racial distribution was 90% Caucasian, 9% Negro, and 1% Oriental, approximating the admission pattern of The New York Hospital. The eighty-four tumors occurred in 83 patients; one patient had a jejunal leiomyoma detected and removed eight years following resection of a similar tumor in the duodenum. The symptoms that attracted clinical attention are given in Table 2. Six of these tumors were discovered as incidental autopsy findings, four of these thought in retrospect to have caused symptoms. The remaining tumors were sufficiently symptomatic to require operation.

Obstruction

Obstruction was the most common symptom in the group as a whole, and was evident in one form or another in 42% (35) of the total. Most often the obstruction was evidenced by crampy intermittent abdominal pain. Often the symptoms abated and allowed study of the patient and elective surgery to be performed. Over half (20) of the patients, however, entered the hospital and had an emergency laparotomy done for unrelieved obstruction. Of these patients 15 had had prior symptoms of intermittent obstruc-

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TABLE 1. *Distribution of Benign Tumors of the Small Intestine*

Histologic Type	Total Number	Location: Duodenum	Jejunum	Ileum
84 New York Hospital Cases				
Leiomyoma	36	7 (19%)	14 (39%)	15 (42%)
Lipoma	22	7 (32%)	3 (14%)	12 (54%)
Angioma	9		3 (33%)	6 (66%)
Adenoma	7	2 (28%)		5 (72%)
Neurofibroma	6	1 (17%)	1 (17%)	4 (66%)
Fibroma	4		1 (25%)	3 (75%)
	84	17 (20%)	22 (26%)	45 (54%)
1721 Collected Cases Where Location was Specified				
Leiomyoma	380	73 (19%)	168 (44%)	139 (37%)
Lipoma	301	72 (24%)	54 (18%)	175 (58%)
Angioma	212	18 (8%)	99 (47%)	95 (45%)
Adenoma	245	97 (39%)	61 (25%)	87 (36%)
Neurofibroma	76	12 (16%)	24 (32%)	40 (52%)
Fibromas	153	8 (5%)	26 (18%)	119 (77%)
Fibromyomas	64	12 (18%)	17 (26%)	35 (56%)
Myofibromas	10	1 (10%)	3 (30%)	6 (60%)
Myxomas	6		1 (16%)	5 (84%)
Villous Adenomas	15	9 (60%)	3 (20%)	3 (20%)
Neurolemmomas	2		1 (50%)	1 (50%)
Polyps	245	61 (25%)	63 (26%)	121 (49%)
Fibroadenomas	12	4 (33%)	2 (17%)	6 (50%)
	1721	367 (21%)	522 (30%)	832 (49%)

tion and only five underwent emergency surgery for obstruction without prior symptoms.

Hemorrhage

Twenty-eight patients (34%) presented with bleeding, in 13 severe enough to require emergency laparotomy. Hemorrhage has been reported in from 9% to 58% of previous series.[2,5,7,10,14,16,19,20,22] Hemorrhage has generally been most common from hemangiomas and leiomyomas; other tumors do cause bleeding but less often.[2,9,14,16]

Pain

A significant number of patients, including most of those noted above to have chronic obstructive symptoms, presented with some pain. In 18 (22%) pain was a predominant symptom. It was always of a colicky nature and usually followed meals. This closely resembles the pain described by others[16,19] although a burning component has at times caused false diagnosis of peptic ulcer disease.[19] The reported incidence of significant pain from benign small bowel tumors ranged from 12% to 36%.[2,5,7,10,14,16,19,20,22]

Weight Loss

Weight loss is an uncommon symptom from small bowel tumors. In our experience only one patient reported preoperative weight loss. This patient had a duodenal tumor and regained his weight following removal. Among the patients observed with acute intestinal obstructions, weight loss may be common especially when the obstruction is high in the small bowel. This loss of weight however is transient and corrected by replacement of fluid losses from vomiting. Other reports except for River's cumulative review also indicate that weight loss is a rare symptom.

Specific Tumors

Leiomyomas. Leiomyomas are intramural tumors composed of whorls of smooth muscle closely resembling the normal muscularis. These tumors are usually well circumscribed but ordinarily lack a true connective tissue capsule. They grow by expansion with compression of lumen

TABLE 2. *Clinical Features of Benign Small Bowel Tumors*

Tumor	Total Cases	Bleeding	Pain	Acute Obstruction: Intusussception	Other	Chronic Obstruction	Weight Loss	Mass
84 New York Hospital Cases								
Leiomyoma	36	14(39%)	6 (17%)	3 (8%)	2 (5%)	11 (30%)		2 (5%)
Lipoma	22	6 (27%)	5 (23%)	7 (31%)	2 (1%)	10 (45%)		
Angioma	9	6 (67%)	3 (33%)			0 (0%)		
Adenoma	7	2 (29%)	1 (14%)	3 (42%)		2 (29%)	1 (12%)	
Neurofibroma	6		2 (33%)	3 (50%)		4 (67%)		
Fibroma	4		1 (25%)		0 (0%)	3 (75%)		0 (0%)
Totals	84	28 (34%)	18 (22%)	16 (19%)	4 (5%)	30 (37%)	1 (1%)	2 (2%)
Collected Cases (1047 tumors of similar cell types in which adequate data is available for tabulation)								
Leiomyoma	196	87 (45%)	106 (54%)	25 (13%)	27 (29%)	57 (29%)	26 (13%)	51 (26%)
Lipoma	224	32 (14%)	35 (16%)	114 (51%)	22 (10%)		21 (9%)	56 (25%)
Angioma	134	55 (41%)	42 (31%)	18 (13%)	20 (15%)		9 (7%)	14 (10%)
Adenoma	232	70 (30%)	50 (25%)	119 (51%)	31 (13%)		39 (17%)	62 (27%)
Neurofibroma	90	32 (36%)	37 (41%)	7 (8%)	20 (22%)		5 (6%)	22 (25%)
Fibroma	163	30 (18%)	116 (69%)	105 (63%)	33 (20%)		32 (19%)	45 (27%)
Totals	1037	306 (30%)	386 (38%)	388 (38%)	183 (18%)		132 (13%)	250 (24%)

and stretching of the mucosa as the mass increases in size. Ulceration is due to compromise of attenuated blood vessels rather than invasion. The bulk of the tumor may be on the serosal side of the bowel and the lesion may have a dumbbell shape due to the increased resistance to expansion by the muscularis as compared to the serosa and submucosal areolar connective tissue.

Some nuclear pleomorphism may be seen in benign tumors. A diagnosis of malignancy is based on increased mitotic activity indicated by more than one mitotic figure per high power field. Palisading of nuclei into rows is frequently seen in leiomyomas and should not be considered characteristic of a nerve sheath tumor which is an extremely rare entity in the intestine. Hyalinization of variable degree may be observed in some leiomyomas which is undoubtedly the basis of designations such as fibromyoma or myofibroma in other reports.

These tumors were the most prevalent of all cell types in both our own and the previously reported experiences. The distribution through the small bowel was rather uniform, if the shorter duodenal length is considered. Both bleeding and pain were frequent presenting symptoms,[3,16] each occurring in over 40% of patients (combining collected cases with our own). Acute obstruction was common, occurring in over one-fourth of patients, but was less often due to intussusception than with some other tumors. Perforation is very rare (seven cases have been reported), but except for one neurofibroma, small bowel tumors that do perforate have been universally leiomyomas.[16] Peak age of occurrence is in the fifth decade[16] and there is no sexual predominance.

Lipomas. Intestinal lipomas are well-circumscribed masses of adult adipose tissue, occasionally surrounded by a delicate collagen capsule. Most of these lesions arise within the submucosa and they grow by expansion with compression of the lumen. It seems likely that many of these lesions are hamartomas rather than neoplasms. Most lipomas have scant vasculature but in some lesions, vessels are more prominent and these are occasionally designated angiolipomas. Fat necrosis may be prominent in some tumors and the presence of foreign body type cells with fat necrosis should not be mistaken for liposarcoma.

Over half of the lipomas reported, in both our own and cumulative experience were in the ileum. There was a high incidence (51% of those in previous reports and 31% of our patients) of intussusception. Nearly half of our patients had symptoms of chronic intermittent obstruction. Bleeding was less frequent than in the other common tumors, being found in only 14% of collected cases. Fifteen of our 22 patients were men. Our peak age of occurrence was the seventh decade, in River's cases more patients were in their sixth decade.

Angiomas. True angiomas are discrete, well circumscribed or encapsulated masses, composed of blood vessels. Capillaries and thin-walled veins usually predominate, but arteries and arterioles may be seen. Some of these lesions are probably hamartomatous arteriovenous malformations rather than vascular neoplasms, but their circumscription distinguishes them from disorders such as Osler-Weber-Render syndrome. The lesions are usually in the submucosa.

Angiomas were the third most common of the tumors in our series, and fourth among collected cases, comprising about 10–12% of each group. Bleeding was the predominant presentation occurring in 6 of our 9 patients, and slightly less often in other reports. Most of the patients were in their seventh decade. There were five men and four women. Pain has been reported in a substantial number of cases and was present in three of our nine cases.

Adenomas. Adenomas are neoplasms of surface epithelium usually in the form of a nodular mass with a stalk composed of normal mucosa and submucosa. The rare epithelial tumors of small bowel with a uniform villous or papillary pattern were associated with invasive carcinoma in all instances in our experience. Some lesions originally classified as adenomas proved to be inflammatory polyps on review, and are not included in our report. Adenomas differ from the polyps seen in Peutz-Jeghers syndrome in that the latter have a smooth muscle component suggesting that they are hamartomas.

There were only 7 adenomas in our series. Adenomas represent 12 percent of all reported cases. When ours and collected reports are combined, there is a fairly even age distribution and acute obstruction, usually by intussusception, was the predominant (64%) presentation. A disproportionate number (39%) were found in the duodenum.

Neurofibromas. Neurofibromas are isolated tumors of nerve sheath origin usually unencapsulated and poorly circumscribed lesions composed of wavy, fibrillary elements. These lesions are identical to those seen in systemic neurofibromatosis (von Recklinghausen's disease). They are found in submucosa, muscularis or serosa, reflecting the distribution of nerve tissue.

There were 6 neurofibromas in our series; they are somewhat less common in total collected cases. They usually occur in the ileum, and bleeding (36%) is more frequent than other symptoms. Intussusception occurred in 3 of our cases but was reported in only 7 of 90 neurofibromas in the cumulative review. Most reported cases have been in their fifth or sixth decade. One instance of perforation has been reported.[16]

Fibromas. Fibromas are circumscribed tumors consisting of dense thick collagen bundles and variable numbers of mature appearing fibroblasts. These tumors may occur in submucosa, muscularis or serosa. In some instances scar tissue may be indistinguishable from a fibroma but usually the fibroma shows a greater degree of circumscription.

Four small intestinal fibromas were found in our material,

all in male patients; three of these were in the ileum. From the collective review it may be seen that 65% of fibromas cause intussusception and obstruction. Pain was the second most common finding. Fibromas are mainly found in adults with evenly distributed age occurrence.

Diagnosis

The diagnosis of small bowel tumors is difficult and most often only established at the operating table. Tumors of the stomach and the large bowel are fairly easily diagnosed by conventional barium x-rays or by direct visualisation through the gastroscope or colonoscope. The large volume of succus entericus, the length of the small bowel and rapid passage of the medium through the intestine tend to make the x-ray diagnosis of small bowel tumors difficult and not easily reproducible. In our cases a radiological diagnosis of small bowel tumor was made 30 times.

The most common symptoms of small bowel tumor, obstruction and bleeding are also quite nonspecific and do not necessarily lead to the correct diagnosis. In the case of small bowel obstruction, the most common cause is intestinal adhesion or hernias. In the case of blood in stools the most common sources are in the colon, anus, stomach or duodenum.

The clinician however should become suspicious when a patient with no previous operation has intermittent bouts of small bowel obstruction, or intermittent crampy abdominal pain. Similarly the patient with occult blood in the stool and normal examination of the upper and lower gastrointestinal tract should be suspect. The sign of frankly bloody stool or currant jelly stool as seen in infarct with intussusception may of course be pathognomic. The clinician who encounters patients of this type must bear the possibility of small bowel tumor in mind, and also recognize that the diagnosis may not be made except through an operation. Intussusception in adults is almost always caused by a tumor, benign or malignant.

Treatment and Results

All of the tumors diagnosed intraoperatively were treated by local segmental or wedge resection. There were no deaths in operation or operative admission. No patient suffered significant postoperative complications.

References

1. Botsford, T. W., Crowe, P. and Crocker, D. W.: Tumors of the Small Intestine. Am. J. Surg.' 103:358, 1962.
2. Braasch, J. W. and Benbo, M. E.: Tumors of the Small Intestine. Surg. Clin. North Am., 44:791, 1964.
3. Cherry, J. W. and Hill. R. L.: Leiomyomas of Jejunum: Neoplasms Imitating Symptoms of Duodenal Ulcer. Arch Surg., 62:580, 1951.
4. Cohen, A., McNeill, A., Tenz, J. J. and Lawrence, W.: Neoplasms of the Small Intestine. Am. J. Dig. Dis., 16:815, 1971.
5. Darling, R. C. and Welch, C. E.:Tumor of the Small Intestine. N. Eng. J. Med., 260:397, 1959.
6. De Castella, H.: Brunner's Gland Adenoma. An Unusual Cause of Internal Bleeding. Br. J. Surg., 53:153, 1966.
7. Ebert, P. A. and Zuidema, G. D.: Primary Tumor of the Small Intestine. Arch. Surg., 91:452, 1965.
8. Eckel, J. M.: Primary Tumors of the Jejunum and Ileum. Surgery, 23:467, 1948.
9. Hancock, R. J.: An 11 year Review of Primary Tumors of the Small Bowel Including the Duodenum. Can. Med Ass. J., 103:1177, 1970.
10. Hashe Mian, M.: Familial Fibromatosis of the Small Intestine. Br. J. Surg., 40:346, 1953.
11. Heurteaux, A.: Note Sur les Tumeurs Benignesde l'intestin. A. Arch Prov chir., 8:701, 1899.
12. Kelsey, J. R., Bentlif, P. S. and Casey, T. M.: Small Bowel Neoplasms: Report of 100 Cases. Tex. Med., 61:383, 1965.
13. Moertel, C. G., Sauer, W. G., Dockerty, M. B. and Baggentoss, A. M.: Life History of the Carcinoid Tumor of the Small Intestine. Cancer, 14:903, 1961.
14. Ostermiller, W., Joergenson, E. J. and Weibel, L.: A Clinical Review of Tumors of the Small Bowel. Am. J. Surg., 111:403, 1966.
15. Raiford, T. S.: Tumors of the Small Intestine. Arch. Surg., 25:122, 1932.
16. River, L., Silverstein, J. and Tope, J. W.: Collective Review: Benign Neoplasms of the Small Intestine. A Critical Comprehensive Review with Reports of 20 New Cases. Int. Abstr. Surg., 102:1, 1956.
17. Sawyer, R. B., Sawyer, K. C., Jr., Sawyer, K. C., Sr. and Lansen, R. R.: Benign and Malignant Tumors of the Small Intestine. Am. Surg., 29:268, 1972.
18. Schatzki, R.: Small Intestine Enema. Am. J. Roentgenol., 50:743, 1943.
19. Schmutzer, K. J., Mulleran, W. M. and Regan, J. F.: Tumors of the Small Bowel. Am. J. Surg., 108:270, 1964.
20. Sladky, F.: Rare Complication of Benign Small Intestine Tumor. Cas op lek csek, 75:1379, 1936.
21. Strinberg, L. S. and Sheiber, W.: Villous Adenomas of the Small Intestine. Surgery, 71:423, 1972.
22. Warshauer, Z. B. and Nelson, R. E.: Neurofibroma of Jejunum with Perforation: Case Report. Am. Surg., 19:467, 1953.
23. Weber, H. M. and Kirklin, B. R.: Roentgenologic Manifestations of Tumors of the Small Intestine. Am. J. Roentgenol., 47:243, 1942.
24. Wiancko, K. B. and MacKenzie, W. C.: Primary Tumors of the Small Bowel and its Mesentery. Can. Med. Ass., 88:1225, 1963.